Case Report
JCAM

A very rare case of sphenoid lymphoma presenting as cavernous sinus syndrome

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Abstract

A 4-year-old boy admitted to our clinic with unilateral ophthalmoplegia, ptosis, and proptosis. He has been suffering the symptoms of a headache, nausea, dyspnea, fatigue, weakness, and loss of appetite for about three months. Complete blood count showed an increased white blood cell count with 77% blast cells, anemia, and thrombocytopenia. Magnetic resonance imaging (MRI) examination revealed the mass was obliterating the sphenoid and cavernous sinuses also causing osseous changes of the sinus walls. We performed endoscopic surgical decompression to relieve the symptoms of cavernous sinus syndrome also to obtain enough tissue samples for pathologic diagnosis. Pathologic examination revealed that the mass was diffuse B cell non-Hodgkin lymphoma and the patient was referred to pediatric oncology department for further treatment.

Keywords

Non-Hodgkin Lymphoma; Cavernous Sinus Syndrome; Ophthalmoplegia

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Introduction

Cavernous sinus syndrome present with the symptoms related to the nerves passing through the cavernous sinus. Painful ophthalmoplegia (cranial nerves III, IV, and VI), proptosis, ocular or conjunctival congestion, Horner syndrome, trigeminal sensory loss, visual loss, and elevation of ocular pressure. Various combinations of these symptoms may be encountered. There are many reasons causing the situation such as infectious, inflammatory, vascular, traumatic, and neoplastic processes. Symptoms are typically unilateral but may be bilateral when there is a neoplastic process [1-3]. Besides, there are some specific situations causing cavernous sinus syndrome including carotid cavernous fistulas, carotid artery aneurysms, tumors, and Tolosa-Hunt syndrome [3].

In this report, we present a very rare case of B cell non-Hodgkin lymphoma which is obliterating the sphenoid sinus and causing cavernous sinus syndrome.

Case report

A 4-year-old child presenting with painful unilateral ophthalmoplegia, ptosis, and proptosis attended to our neurosurgical department. He has been suffering the symptoms of a headache, nausea, dyspnea, fatigue, weakness, and loss of appetite for about three months. He was referred to the radiology department for possible intracranial mass, and MRI examination was performed to rule out any intracranial masses. MRI investigation revealed the mass which was totally obliterating the sphenoid sinuses. T1 weighted images showed that the mass was contingent with ethmoid sinuses anteriorly, nasopharynx inferiorly and cerebral gyruses superiorly but not invading them. The mass consisted of hyperdense areas suggesting hemorrhage or calcification (Figure 1). The mass was hypodense on T2 weighed images consistent with MRI findings of lymphomas (Figure 2). Clivus, nasopharynx and sellar region were free of the disease. Vascular supply of the mass was weak on MRI angiography (Figure 3). The patient was referred to our clinic for evaluation of possible sphenoid mass etiologies. We performed ENT examination which was completely normal. On the third day, the patient developed sudden and complete visual loss. Despite conservative therapy with a high intravenous dose of methylprednisolone, we decided to operate the patient for absolute tissue diagnosis and surgical decompression. Under general anesthesia, the patient was operated endoscopically with 0°and 30° endoscopes. The mass was observed in left sphenoethmoid recess, and multiple punch biopsies were obtained for tissue diagnosis. Frozen section samples were reported as large round cell tumor which was non-specific. We decided to complete the decompression procedure. The tumor was discharged from sphenoid and cavernous sinuses (Figure 4). Patient's symptoms relieved immediately after the surgery. The final histological diagnosis was diffuse large B-cell type non-Hodgkin lymphoma. The patient was referred to the Medical Oncology department and received chemotherapy of cyclophosphamide, hydroxydaunorubicin, oncovin and prednisone combined with 40 Gy radiotherapy.

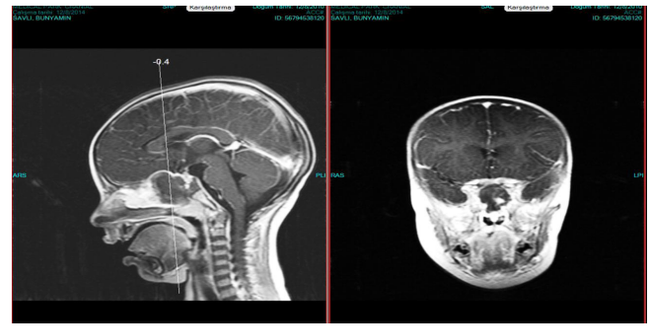

Figure 1. T1 weighted images showing hyperdense areas

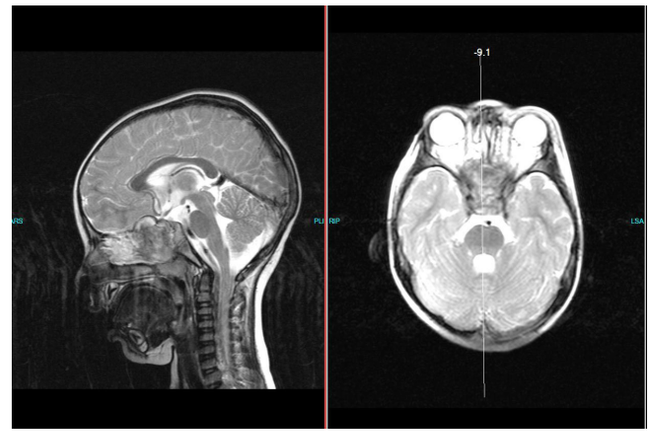

Figure 2. T2 weighed images

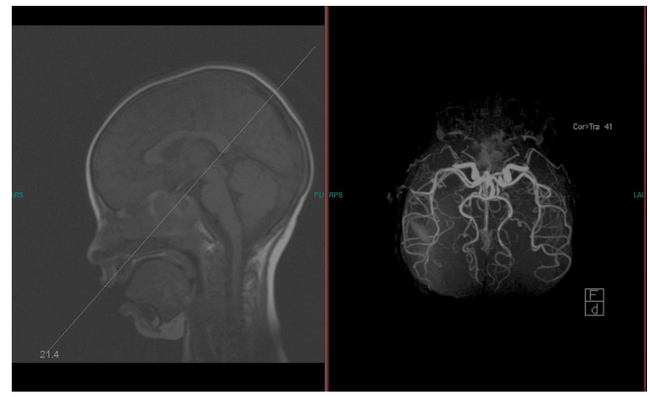

Figure 3. MR Angiography appearance

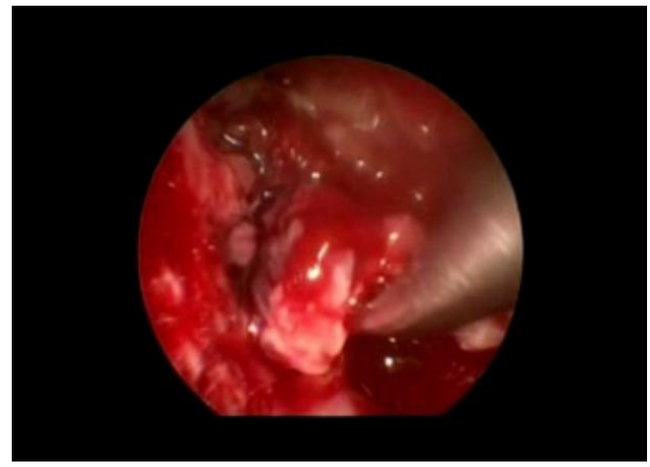

Figure 4. Intraoperative appearance

Discussion

Sphenoid sinus tumors are very rare and seldom associated with symptoms such as nasal discharge and obstruction or epistaxis. A headache and cranial neuropathies are the most common presenting symptoms [4]. Because of the proximity, tumors of the sphenoid bone may invade many important skull base structures. Levine pointed out 13 critical structures that are at risk when there is a sphenoid disease: the dura, the internal carotid artery, the cavernous sinus, the optic nerve, the abducens nerve, the oculomotor nerve, the pituitary gland, the

trochlear nerve, the ophthalmic nerve, the maxillary division of the trigeminal nerve, the sphenopalatine ganglion and artery, and the pterygoid canal and nerve [5].

Paranasal lymphomas are mostly affecting elderly patients, with a median age of 5 to 60 years [6]. However, there are a few reported non-Hodgkin lymphoma cases affecting children [3].

Also, there are several reported cases in adults in which cavernous sinus syndrome occurred as a manifestation of systemic non-Hodgkin lymphoma [7]. The literature on children is more scarce [8]. Diagnosis of paranasal lymphoma is usually delayed because the tumors in this area appear with a very few symptoms and are usually at an advanced stage at the time of diagnosis.

The cavernous sinuses are paired venous structures located on both sides of the sella turcica. Multiple important structures such as the carotid artery; its sympathetic plexus; cranial nerves III, IV, and VI; and the ophthalmic branch of cranial nerve V pass through the sinuses. Cavernous sinus syndrome caused by a tumor and acute or slowly progressive ophthalmoplegia is the dominant presentation, with diplopia the most common symptom. Cavernous sinus tumors are the predominant cause of cavernous sinus syndrome [8]. Tumors may be primary, locally spreading, or metastatic [9]. Meningiomas and neurofibromas are the most common primary tumors [9]. Locally spreading tumors are nasopharyngeal and pituitary originating tumors. Primary tumors are the most common neoplasms causing cavernous sinus syndrome. Akinci et al. reported a cavernous sinus syndrome as the initial presentation of childhood non-Hodgkin lymphoma [3].

This report indicates that cavernous sinus syndrome may occur as the initial presentation of non-Hodgkin lymphoma in children, and endoscopic tissue sampling and decompression surgery should be done immediately.

Scientific Responsibility Statement

The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.

Animal and human rights statement

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this article.

Conflict of interest

None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.

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